

## PHACOEMULSIFICATION IN PSEUDOEXFOLIATION (PEX) SYNDROME

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**SUMMARY** – The aim is to present the phacoemulsification phaco-chop technique in a patient with pseudoexfoliation (PEX) syndrome without the use of additional pupil dilatation methods. Phacoemulsification surgery and posterior chamber intraocular lens implantation in patients with PEX syndrome is associated with a higher rate of intraoperative complications such as zonular dehiscence, capsular rupture, vitreous loss and dropped nucleus. Many options are available for pupils that cannot be dilated sufficiently with pharmacologic agents, such as viscodilatation (Healon 5), bimanual stretching, iris retractor-hooks, and many others. We present cataract surgery in a patient with sufficient mydriasis despite PEX syndrome. The operation was done successfully without intraoperative and postoperative complications. It is concluded that cataract surgery in patients with PEX syndrome is more complicated because of zonular weakness and poor pupillary dilatation. Therefore, these patients should be managed with utmost care and operated on in time by an experienced surgeon.

**Key words:** *Phacoemulsification – methods; Exfoliation syndrome – complications; Cataract – surgery*

### Introduction

Pseudoexfoliation (PEX) syndrome is a generalized disorder of extracellular matrix characterized by the production and progressive accumulation of fibrillar material in all structures of the anterior segment of the eye (palpebral and bulbar conjunctiva, corneal endothelium, angle of the anterior chamber, iris, muscle dilatator and blood vessels of the iris, epithelium of the ciliary body, zonules, anterior lens capsule and anterior vitreous surface) and also in connective tissue portions of various visceral organs<sup>1-3</sup>.

Cataract surgery in patients with PEX syndrome is associated with many complications because of insufficient mydriasis, weakened zonular attachments and lens instability. Moreover, many patients with PEX syndrome have a pseudoexfoliation glaucoma which substantially increases the risk of surgery.

PEX syndrome leads to an increased risk during cataract surgery of zonular dialysis, rupture of the posterior capsule and luxation of the lens into the vitreous as a consequence of insufficient zonules<sup>4</sup>. Furthermore, poor pupillary dilatation, which is a frequent manifestation of PEX, increases the risk of these complications. Many surgical options are available for pupils that cannot be dilated sufficiently with pharmacologic agents<sup>5,6</sup>. Sphincterectomy and other incisional methods were popular in the past, but many surgeons have stopped using these techniques because of postoperative issues with cosmetically and functionally unacceptable iris deformities. The use of iris-retractor hooks, stretch pupilloplasty techniques, and pupil-dilatator rings enable adequate pupil dilatation without severe iris deformities<sup>5,7,8</sup>.

Many studies have shown that phacoemulsification in cataract treatment is an efficient and safe procedure with improvement of the patient's visual acuity and a low number of postoperative complications even in risk patients such as those with PEX syndrome<sup>4,9,10</sup>.

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## Patient and Methods

We present the phacoemulsification phaco-chop technique in a patient with PEX syndrome without the use of additional pupil dilatation methods. Our patient was a 73-year-old man.

Preoperatively, pupils were dilated with phenylephrine hydrochloride 10%, tropicamide 1% eye drops, 3 times every 10 minutes starting 1 hour before the surgery. Tetracaine eye drops were given 4-5 times within a 15-min interval just before the surgery. A limbal self-sealing incision was made with a 3.0 slit knit superotemporally on the right eye and superonasally on the left eye. Then, sodium hyaluronate 3% (Vitrax II, AMO) was injected into the anterior chamber. The second incision was made in clear cornea. The pupil size measured just before anterior capsulorhexis was initiated was 6 mm. Continuous curvilinear capsulorhexis was done with a needle and capsulorhexis forceps. Cortical cleaving hydrodissection and rotation was done with a small cannula. It is important to perform it very carefully due to zonular instability.

Phacoemulsification was performed by use of the Universal II phacoemulsifier (Alcon). The first step of phaco was an initial crack (chop *in situ*) performed by phaco tip (ABS Cobra microtip) and chopper (which moves from periphery to the center). Then, the hemi-nucleus was divided mechanically very carefully into multiple small fragments using the phaco chop technique. The nuclear fragments were consumed within the central space in the pupillary plane.

Typical machine settings during chopping were US power 60-70%, vacuum 250-400 mm Hg and aspiration flow rate (AFR) 20-24 cc/min. Vacuum was progressively reduced as the chopped fragments were removed and during the removal of the last fragments it was 100 mm Hg.

The epinucleus was removed with a phaco probe and the machine parameters during the procedure were US power 60-70%, vacuum 250-400 mm Hg and aspiration flow rate 16 cc/min. After removal of the epinucleus, the cortex was aspirated with an automated irrigation/aspiration (I/A) probe. Then the anterior chamber and capsular bag were filled with a viscoelastic agent (Vitrax II, AMO). The foldable silicone (PhacoFlex II, AMO) intraocular lens (IOL) was implanted very gently in the capsular bag using the Unfolder. The viscoelastic agent was then removed with an I/A probe.

Postoperative medication comprised of neomycin, polymyxin B and dexamethasone (Maxitrol) eye drops 4 times a day and ointment once a day for 1 month.

## Discussion and Conclusion

Poor pupillary dilatation is a frequent manifestation of PEX syndrome, and these patients often have loss of zonular integrity as well. Because of zonular problems, phacoemulsification surgery in eyes with PEX syndrome is associated with a higher rate of intraoperative complications such as zonular dehiscence, capsular rupture, vitreous loss and dropped nucleus<sup>11</sup>. Poor pupillary dilatation further increases the risk of these complications<sup>7</sup>.

Some studies conclude that the increased rate of intraoperative complications during cataract extraction in patients with PEX syndrome stems from zonular weakness rather than capsule tear<sup>12</sup>. Most problems related to poor zonular support of the lens can be avoided with the use of specialized surgical techniques that do not tear the remaining zonules. Implantation of endocapsular rings increases the stability of the capsular bag and prevents further damage to the zonules<sup>12</sup>. Endocapsular ring implantation is recommended when the presence of phacodonesis occurs due to severe loss of zonular integrity. Sometimes we stabilize the capsular bag using iris dilators by holding on to the rim of the anterior capsulorhexis.

Mechanical dilatation of the small pupil enables phacoemulsification to perform safely in eyes with PEX syndrome and small pupils. Most of the small pupil widening techniques provide sufficient dilatation with adequate visualization, reduce the risk of intraoperative complications and allow the surgeon to perform phacoemulsification safely.

The pupil size necessary for safe phacoemulsification in eyes with PEX syndrome depends on many factors such as the surgeon's level of experience and the nature of the cataract. As a general rule, a pupil size large enough for adequate capsulorhexis is considered large enough to perform surgery<sup>11</sup>. A 5.0 to 6.0 mm diameter of capsulorhexis is adequate for routine cases, but in patients with PEX syndrome a larger capsulorhexis size is often preferred.

In our case, the patient had sufficient mydriasis despite the PEX syndrome and mechanical dilatation was not necessary. All steps of the surgery had to be performed with extreme care and caution due to marked zonular instability. Three months after the surgery, the patient's corrected visual acuity was 1.0.

In conclusion, the modern cataract surgery by phacoemulsification can be performed safely in patients with PEX syndrome by an experienced surgeon operating the patient on time and using special care and caution, in spite of the high risks of the possible intraoperative and postoperative complications.

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## Sažetak

### FAKOEMULZIFIKACIJA KOD SINDROMA PSEUDOEKSFOLIJACIJE (PEX)

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Cilj je prikazati tehniku fakoemulzifikacije phaco-chop kod bolesnika sa sindromom pseudoeksfolijacije (PEX) bez primjene dodatnih metoda širenja zjenice. Fakoemulzifikacija i implantacija intraokularne leće u stražnju sobicu kod bolesnika sa sindromom PEX je povezana s povećanom učestalošću intraoperacijskih komplikacija, kao što su dehiscencija zonula, ruptura kapsule, gubitak staklovine te utonuće nukleusa leće. Za zjenice koje ne pokazuju zadovoljavajuće širenje uz primjenu farmakoloških sredstava postoje brojne druge metode kao što su viskodilatacija (Healon 5), bimanualno 'istezanje', 'retraktorkukice za šarenicu' te mnoge druge. Opisuje se ovaj zahvat kod bolesnika sa zadovoljavajućom midrijazom usprkos sindromu PEX. Operacija je napravljena uspješno bez intraoperacijskih i poslijeoperacijskih komplikacija. Zaključak je kako je kirurgija katarakte kod bolesnika sa sindromom PEX složenija zbog slabosti zonula i slabo proširene zjenice, zbog čega ovi bolesnici zahtijevaju osobitu pozornost i iskustvo kirurga.

Ključne riječi: *Fakoemulzifikacija – metode; Katarakta – kirurgija; Sindrom eksofolijacije – dijagnostika*

